

Rheumatic constrictive pericarditis

A case report and review of the literature

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Summary

A case of calcific constrictive pericarditis in a young White woman with a convincing history of previous acute rheumatic fever complicated by a possible valvular lesion is presented. Cardiac catheterization confirmed the suspicion of significant cardiac compression. Successful pericardiectomy was carried out, but microscopical examination of the excised pericardium failed to demonstrate a cause. In view of the strong past history of acute rheumatic fever and mild mitral insufficiency demonstrated at cardiac catheterization, the author proposes that the calcific constrictive pericarditis was of rheumatic origin. A review of the literature on the association between rheumatic infection and constrictive pericarditis follows.

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Case report

The patient was a 30-year-old White female schoolteacher with a 7-month history of swelling of the face and ankles, increasing abdominal girth, weight gain, and shortness of breath on moderate exertion. She had also noted occasional palpitations on exercise. There was no history of orthopnoea, paroxysmal cardiac dyspnoea or chest pain.

At the age of 7 years she had apparently been extremely ill with joint pains, fever and chorea and she had been found to have a cardiac murmur. Acute rheumatic fever with heart involvement was diagnosed and she was given prophylactic penicillin orally for some 10 years. During this period she was examined regularly and was told she had a 'leaking heart'. She continued to be asymptomatic and no other medication was deemed necessary.

At 23 years of age she had been delivered of her first child who died because of prematurity but she was apparently quite healthy during her pregnancy. Three years later she underwent repair of uterine prolapse and was noted to have uterine polyps. There was no history of tuberculosis or contact with this disease, and no recurrence of acute rheumatic fever.

On examination at the Cardiac Clinic, Tygerberg Hospital, she appeared to be in good general physical condition apart from moderate bilateral pedal oedema. A 2 cm, slightly tender, firm hepatomegaly and some ascites was found, but there was no splenomegaly. Bilateral basal crepitations were also present posteriorly.

Examination of the cardiovascular system revealed a radial pulse somewhat small in volume, regular, with all the peripheral

pulses equally palpable, while no radiofemoral delay could be detected. The blood pressure was 100/70 mmHg, with no definite pulsus paradoxus. The jugular venous pressure was elevated to 10 cm with easily visible 'a' and 'v' waves exhibiting rapid 'x' and 'y' descents and a positive Kussmaul sign. The cardiac apex was located at the fifth intercostal space in the midclavicular line and appeared normal. A mild diastolic left parasternal heave with no epigastric pulsation was felt. Over the mitral area the first and second heart sounds were normal, but there was an adventitious sound in early diastole suggestive of a 'pericardial knock'. A soft grade 1/6 pansystolic murmur, not varying with respiration and with very little radiation, was audible. There was no apical diastolic murmur. At the pulmonary area an abrupt and short-lived wide splitting of the second heart sound at the onset of inspiration was heard. There were no adventitious sounds or murmurs at the base of the heart. The remainder of the clinical examination was negative.

A resting ECG (Fig. 1) showed a sinus rhythm of 70/min, a PR interval of 0.16 seconds, and a mean frontal QRS axis of 85°. There was no evidence of atrial or ventricular hypertrophy. Asymmetrical T-wave inversion was present, and the ST segments were horizontal in all the leads.

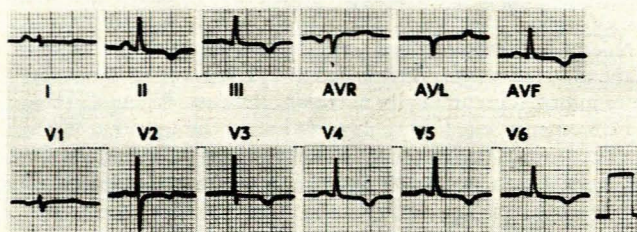


Fig. 1. Resting ECG showing diffuse asymmetrical T-wave inversion and horizontal ST segments.

The chest radiograph (Fig. 2) showed a 'mitralized' heart with possible right ventricular cardiomegaly, a prominent main pulmonary artery segment and left atrial appendage. Extensive dense calcification was seen to involve the pericardium around the coronary sulcus, the diaphragmatic surface, the sternal aspect of the right ventricle, as well as part of the left ventricle. The lung fields appeared normal.

Serum electrolyte and fasting glucose levels, liver function, renal function and haematological investigations including ESR were all normal. Immunological investigations to exclude possible systemic disease were all negative. Serum electrophoresis and immunophoresis showed normal patterns. The Mantoux reaction was negative, as were tests for tuberculosis. An intravenous pyelogram was normal. Echocardiography was not performed.

In view of the clinical diagnosis of significant constrictive pericarditis it was decided to perform cardiac catheterization with a view to corrective surgery.

Cardiac catheterization

This was performed using the standard percutaneous Seldinger technique via the right femoral vein and artery. A 7F

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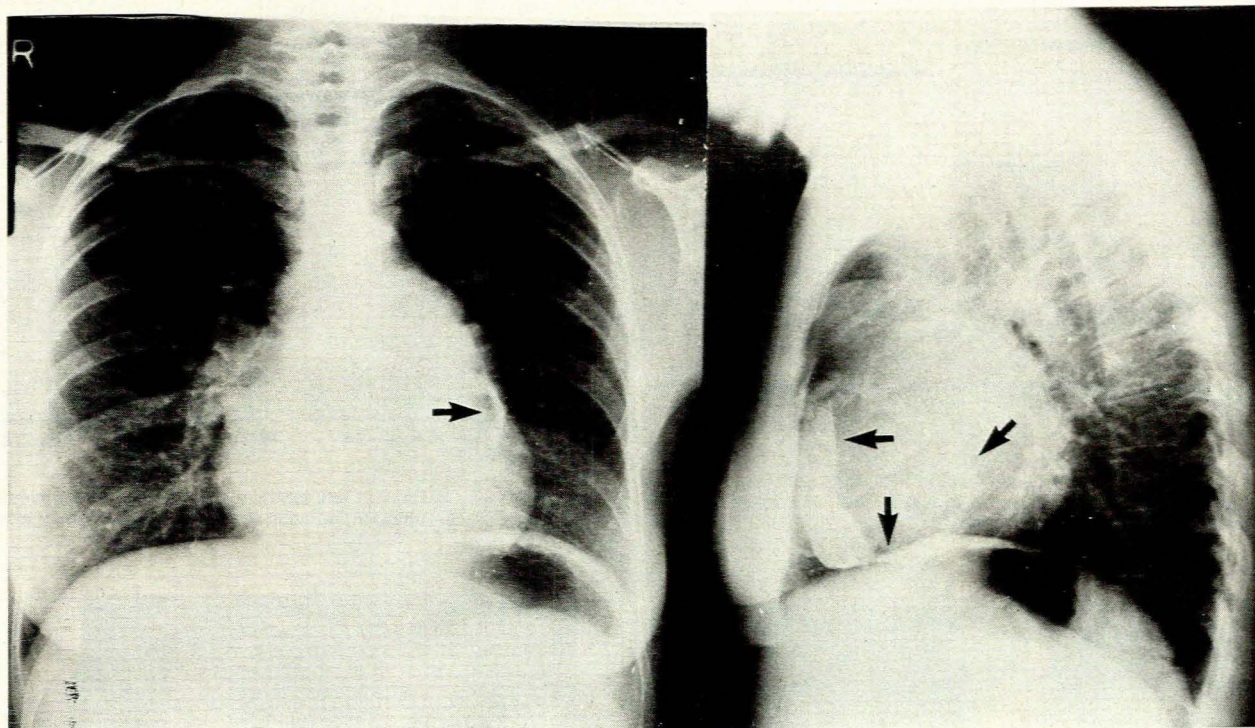


Fig. 2. Chest radiograph (postero-anterior and left lateral views) showing mitralization of cardiac shadow, as well as dense calcification (arrowed). The lung fields are normal.

Goodale-Lubin and pigtail catheter was used to measure intracardiac pressures on the right and left sides, as well as oxygen saturations in the main pulmonary artery and central aorta (Tables I and II). The pressure curve tracings of the right atrium, right ventricle, pulmonary capillary wedge (indirect left atrium), and left ventricle are illustrated in Figs 3 and 4. Pressures in the right atrium and left atrium (pulmonary capillary wedge) were the same because diastolic expansion was markedly restricted by the rigid pericardium. For the same reasons, the diastolic pressures in the right and left ventricles, as well as the main pulmonary artery, were almost identical to the mean pressures in the two atria. The classic prominent 'a' and 'v' waves with marked 'x' and 'y' descents, giving rise to the typical 'M' or 'W' wave forms, can also be seen. Ventricular pressure tracings showed the presence of the 'dip and plateau' pattern resembling a square-root sign. The classic 'ha' waves were also seen during ventricular diastole.

Left ventricular cine angiography (right anterior oblique view) showed a well-contracting ventricle with a mid-cavity deformity due to extrinsic pericardial constriction and a mildly

TABLE II. HAEMODYNAMIC CALCULATIONS

	Result
Cardiac output (Fick) (l/min)	5,6
Cardiac index (Fick) (l/min/m ²)	3,4
LV dp/dt (mm/s)	1 248
Pulmonary vascular resistance (U)	1,1
Systemic vascular resistance (U)	12,1
Pulmonary/systemic resistance ratio	9%

insufficient, non-calcified mitral valve (Fig. 5, left). Right ventricular cine angiography (anteroposterior and left lateral views) indicated extrinsic obstruction of the outflow tract giving rise to a dilated main pulmonary artery. The right ventricle was also hypertrophied with diastolic overload (Fig. 6, left). A right atrial cine angiogram (anteroposterior view) showed a significantly dilated and distorted chamber as well as a dilated superior vena cava. Calcification was clearly evident (Fig. 6,

TABLE I. INTRACARDIAC PRESSURES

Catheter position	Pressure (mmHg)	Comment
Right atrium	'a' wave 22, 'x' descent 18, 'v' wave 20, 'y' descent 17, mean 22	Marked right atrial hypertension with 'W' or 'M'-shaped configuration
Right ventricle	42/15 - 24	Early 'diastolic dip', late 'diastolic plateau', raised diastolic pressure
Main pulmonary artery	42/25, mean 27	Mild pulmonary hypertension, no pulmonary stenosis
Pulmonary capillary wedge	'a' wave 28, 'x' descent 19, 'v' wave 26, 'y' descent 18, mean 22	Moderately elevated pressures, no mitral stenosis
Left ventricle	104/8—24	Elevated end-diastolic pressure; early 'diastolic dip', late 'diastolic plateau'
Ascending aorta	104/74, mean 88	Normal pressures, no aortic stenosis

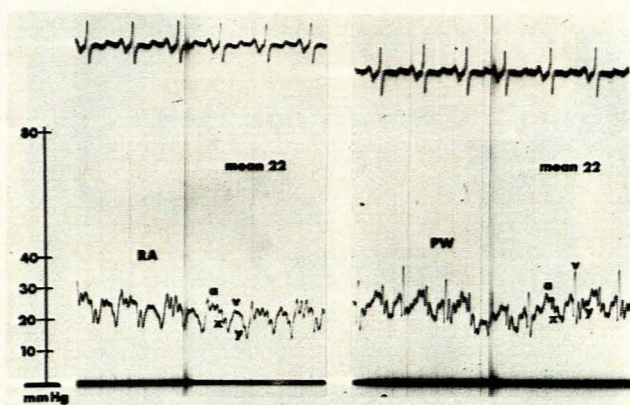


Fig. 3. Pressure tracings of right atrium (RA) and pulmonary capillary wedge (PW). Prominent 'a' and 'v' waves, as well as marked 'x' and 'y' descents are demonstrated (see text).

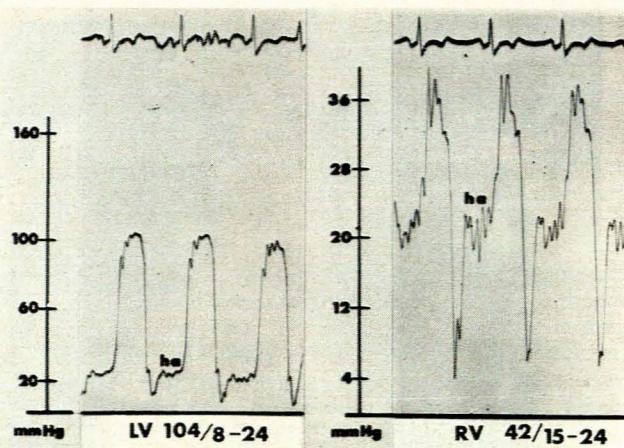


Fig. 4. Pressure tracings of left ventricle (LV) and right ventricle (RV), showing typical diastolic configuration seen with 'ha' waves (refer to text).

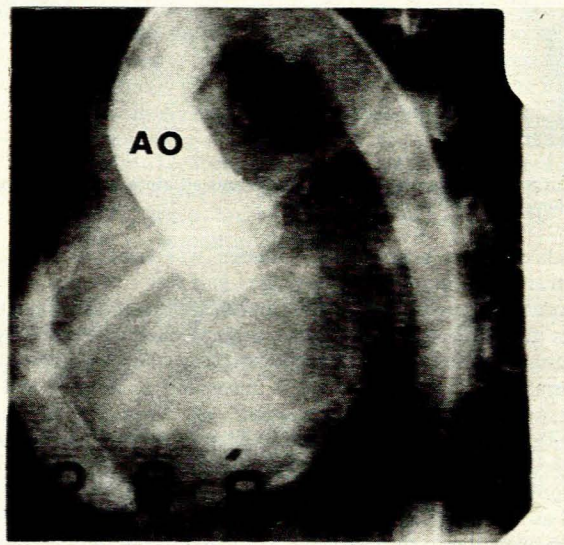
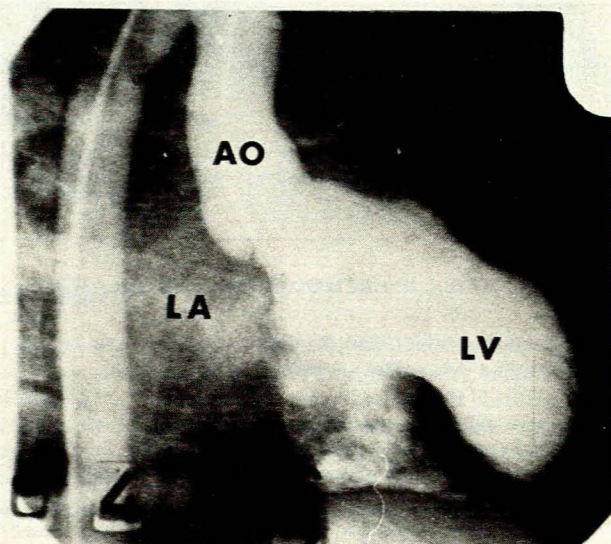


Fig. 5. Left: left ventricular cine angiogram (right anterior oblique view), showing mid-cavity obstruction of left ventricle (LV) due to calcific extrinsic pericardial constriction, as well as mitral insufficiency (LA = left atrium; AO = aorta); right: aortic cine angiogram (left anterior oblique view), showing heavy pericardial calcification, as well as normal aortic valve.

right). Cine angiography of the ascending aorta (left anterior oblique view) showed a normal aortic valve and ascending aorta (Fig. 5, right). The catheterization procedure was completed without any complications.

The severity of the constrictive pericarditis delineated by the cardiac catheterization called for surgery.

Operative findings and surgical correction

A median sternotomy was carried out. The pleura was very heavily calcified over the right atrium in the atrioventricular groove, the inferior surface and the right ventricular outflow tract, as well as over the posterior aspect of the left atrium and appendage. The pericardium adhered to the left ventricle but was not very thickened. A total pericardiectomy was performed with release of the entire left ventricle, right ventricle and right atrium in succession. A small area of the right ventricular outflow tract muscle was also calcified due to extension from the pericardium itself. The operation was successful and the patient was kept on a small dose of diuretic for about 3 months postoperatively.

Histological findings

The pericardium was grossly thickened with extensive fibrosis. Areas of dense calcification were present within the avascular fibrotic tissue, but there was no evidence of inflammatory cells pointing to a definite aetiological factor. There were a few patches of myocardial cells intermingled with fibrotic and calcified areas, verifying the extension of the pathological process from the pericardium to the myocardium.

Follow-up examination

The patient was seen regularly at the Cardiac Clinic for about 2 years after the operation. She denied any symptoms and there was no evidence of cardiac compression clinically. Several repeat ECGs failed to show any change from the pre-operative tracing.

Discussion

Definition and pathology

Clinical constrictive pericarditis is essentially a mechanical

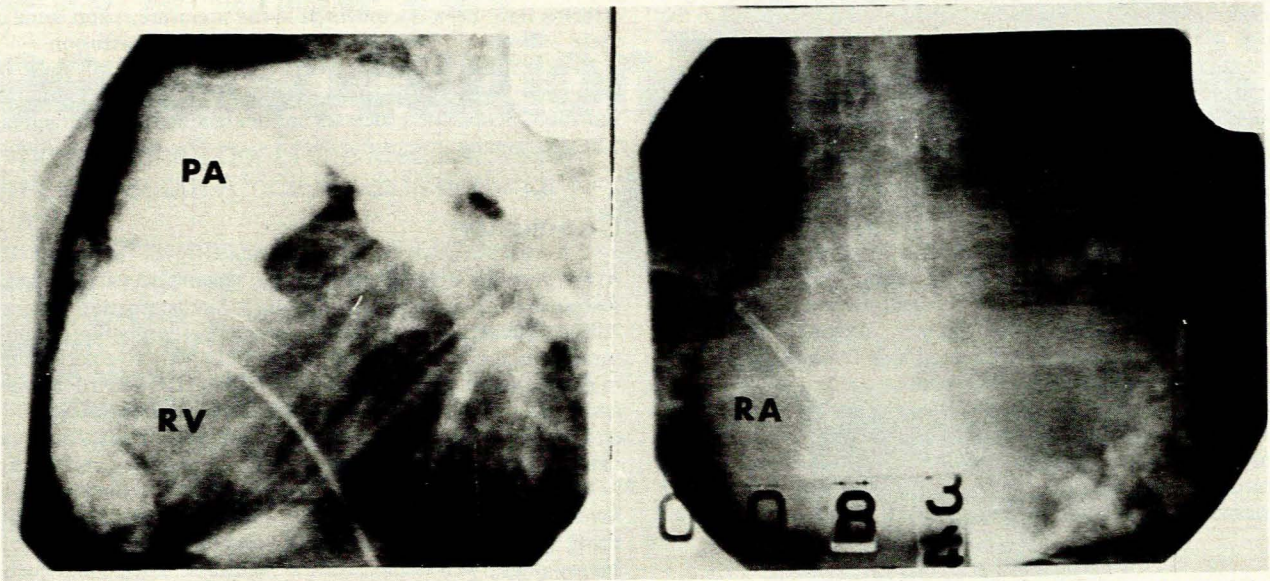


Fig. 6. Left: right ventricular cine angiogram (left lateral projection), showing extrinsic obstruction of right ventricular (RV) outflow tract and dilated main pulmonary artery (PA); right: catheter in right atrium showing extensive calcification.

condition in which the two layers (visceral and parietal pericardium) are adherent as well as thickened, leading to interference with some of the normal functions of the heart, particularly the diastolic filling phase. However, it must be remembered that anatomical pericardial adhesion does not necessarily give rise to clinical pericardial constriction. Calcification of the pericardium occurs in approximately 50% of all reported cases, but is present in over 70% of chronic cases and rarely seen in acute constriction. Extensive calcification of the heart can be present in the absence of constriction.¹

There is little doubt that systolic ventricular function is often altered, but this component is not usually accepted as being an essential feature. Several workers² have demonstrated myocardial fibre atrophy and fibrosis consequent upon chronic pericardial constriction, although the degree of myocardial alteration was not directly related to the pericardial thickness or duration of symptoms.³

Moschcowitz⁴ studied 130 cases of adherent pericardium at autopsy; in 71 cases the pericardium was found to be completely adherent. In these 71 cases a rheumatic aetiology was considered in 53,5%, coronary thrombosis in 8,5%, tuberculosis in 4,2% and polyserositis in 5,6%; there was no apparent cause in 26,8% and 1,4% were due to miscellaneous causes. In 7 of the 130 patients there was an anatomically constrictive pericardium, but only 4 of these had features of clinical constrictive pericarditis (1 with definite past rheumatic fever and rheumatic mitral and aortic valvulitis). The remaining 3 patients were thought to be in the 'early phase of clinical constriction'.

The exact reasons why some patients with rheumatic pericarditis develop pericardial constriction is not known. This may depend upon the degree of severity of the initial acute fibrinous pericarditis, and the duration, as well as the host's immunological response to this inflammation.

Histologically, the pericardium does not show any features of inflammation, but rather the results of it. There is a varying degree of avascular tissue consisting of fibrous tissue which has undergone partial or complete hyalinization. Areas of calcification may be present which vary in magnitude. This makes an accurate aetiological diagnosis difficult in the majority of cases.

Relationship between rheumatic fever and constrictive pericarditis

Much controversy surrounds the development of constrictive pericarditis secondary to acute rheumatic fever; it is generally accepted that cardiac constriction does not occur during the acute infection.

Bland and Jones⁵ performed autopsies on 306 patients who had died of rheumatic fever; they found evidence of acute pericardial inflammation in 55% and acute or chronic pericardial involvement in 80% of cases. They commented that in 'spite of the high incidence of pericardial involvement with rheumatic fever no instance of Pick's syndrome has yet been encountered'. Armstrong⁶ supported their statement in reiterating the 'widely held clinical view that cardiac compression never follows a rheumatic pericarditis. The end-results of rheumatic inflammation are never such as have been described in the constrictive group.' King⁷ went so far as to say that a history of rheumatic infection should make one sceptical about the diagnosis of constriction of the heart.

In 1948 Mortensen and Warburg⁸ analysed 25 cases of constrictive pericarditis and found that only 2 patients had had rheumatic fever. They decided that the association of these two conditions was purely coincidental. Likewise, Jackson⁹ reported a 52-year-old male with constrictive pericarditis who had had three attacks of acute rheumatic fever, evidence of mitral valve stenosis, and no tuberculosis contact. He concluded that tuberculosis was the cause and that rheumatic fever was a chance association.

Elkeles¹⁰ took the opposing view. He described 2 patients with rheumatic mitral stenosis and severe calcific constrictive pericarditis. His comment was that 'rheumatic fever can be an aetiological factor in the production of chronic adherent calcified pericarditis' and that 'the widely held view that tuberculosis is the most common aetiological factor in chronic adherent pericarditis has to be revised'. Chambliss *et al.*¹¹ reported 10 cases of chronic constrictive pericarditis in patients who had had rheumatic fever. They believed that this infection was possibly the cause in only 1 case and that the association was almost certainly coincidental. White¹² corroborated the view of many previous workers in stating that 'constrictive pericarditis following acute rheumatic fever probably does not occur'.

Kaltman *et al.*¹³ described 5 patients who had rheumatic valve disease and aortic insufficiency. Autopsy showed calcified

pericardium in 1, but fibrotic and thickened pericardium in the other 4 patients. He also reported 3 other cases in which rheumatic fever had recurred without ensuing valvular disease, but in which the pericardium was grossly thickened, fibrotic and heavily calcified. Despite these findings the authors went on to state that the rheumatic infection was not a 'causal relationship but a coexisting one'.

Winters and Soloff¹⁴ commented on the fact that rheumatic fever causes pericardial damage which leads to impaired pericardial absorption, thus making chronic pericarditis possible, with the likelihood of pericardial constriction. This concept was further supported by Bedford,¹⁵ who noted that rheumatic fever could cause 'chronic idiopathic pericardial effusion'. Brown¹⁶ also documented this condition following upon acute rheumatic fever with chorea.

Despite the above evidence, the controversy surrounding rheumatic fever as a cause of chronic constrictive pericarditis continues. After reviewing the literature, I feel justified in recording this case as one of chronic calcific constrictive pericarditis directly caused by past acute rheumatic infection.

Differential diagnosis of rheumatic constrictive pericarditis

In South Africa the most common cause of constrictive pericarditis has been accepted as tuberculosis, the majority of cases being in the Black population group. Schrire¹⁷ reviewed 160 cases of pericarditis at Groote Schuur Hospital and found that tuberculosis was responsible for some 50%, and that in a further 25% it was the probable cause. Some 60% of the patients with tuberculous pericardial effusions went on to develop constrictive pericarditis within 6 weeks to a few months, the majority requiring surgery. Desai¹⁸ described 100 Black patients with presumed tuberculous pericarditis, 82% presenting with a pericardial effusion and 18% with constrictive pericarditis. Furthermore, 15 of the 82 patients who initially presented with pericardial effusion developed 'constricting pericarditis' within 4 months.

In Western countries, where tuberculosis is on the decline, this infection is a rare cause of constrictive pericarditis. Bacterial infections caused by staphylococci and pneumococci as well as viral agents (especially coxsackie B) are fairly common causes.¹⁹ Among the parasitic diseases in South Africa, amoebic liver abscess perforating into the pericardium,²⁰ rupture of an echinococcal cyst into the pericardium,²¹ and filariasis²² are known to give rise to constrictive pericarditis. Fungal infections are even more rarely the cause of this condition.²³

The connective tissue disorders have to be considered in the differential diagnosis of rheumatic constrictive pericarditis. Systemic lupus erythematosus commonly causes pericardial effusion, but this rarely develops into constrictive pericarditis.²⁴ Polyarteritis nodosa has also been described as a cause.²⁵ Some 46 cases of constrictive pericarditis secondary to rheumatoid

arthritis have been documented in the literature,²⁶ and some of these patients also had an associated pericardial effusion.

Less common causes of constrictive pericarditis must also be considered in the differential diagnosis. These include trauma, radiation, neoplastic processes and metabolic causes (uraemia). There is usually little difficulty in excluding these conditions.

Conclusion

Acute rheumatic fever and chronic rheumatic valvular heart disease are very common in the non-White population of South Africa. Tuberculosis is widely accepted as probably the only significant aetiological factor in chronic constrictive pericarditis in all population groups in South Africa and the underdeveloped countries. Our patient was interesting in that she was White with an undoubted past history of rheumatic fever. The difficulty in making an aetiological diagnosis in chronic constrictive pericarditis is well known, so that most workers automatically invoke tuberculosis as the cause. If more patients presented with acute pericardial constriction of unknown cause, pericardial biopsy might well demonstrate a rheumatic disease and enable this to be accepted as a cause of chronic constrictive pericarditis. Some clarification of this long-standing clinical problem is attempted in this publication.

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